Specialty Conference

Participants

MARTIN STEIN, MD,
RICHARD BUCHTA, MD
ANDRE RASZYNSKI, MD
WILLIAM L. NYHAN, MD, PhD

From the Department of Pediatrics, University of California Medical Center, San Diego, and University of California, San Diego, School of Medicine.

Refer to: Stein M, Buchta R, Raszynski A, et al: Intussusception—University of California, San Diego, School of Medicine, and the University of California Medical Center, San Diego (Specialty Conference). West J Med 130:35-42, Jan 1979

Intussusception

MARTIN STEIN, MD:* Pediatricians are confronted frequently with infants and toddlers who present with the acute onset of vomiting and abdominal pain. Experienced clinicians are aware that, while gastroenteritis, acute appendicitis, respiratory inflammatory processes, poisoning and urinary tract infections most often account for these symptoms, various forms of intestinal obstruction, though uncommon in young children, must be diagnosed immediately. In particular, intussusception has a reputation for its protean presentation and all-too-frequent delay in diagnosis.

Intussusception is defined as the invagination of one portion of the intestine into another in the direction of peristalsis. The Latin *intus* means "within" and *suscipere* means "to take up." The most common type is ileocolic; less common forms include ileal-ileal, colic-colic and ileal-ileal-colic. The development of intussusception in children is characteristically a spontaneous event during infancy and occurs in previously healthy, well-nourished patients. Two thirds of patients are between 3 and 12 months of age. Less commonly, it may occur in older children, either spontaneously or in association with a leading point—that is, an anatomic lesion that initiates the intestinal invagination process. Known leading points include enlarged mesenteric nodes, hypertrophied Peyer† patches in the distal ileum, Meckel diverticulum, polyp, enteric cyst, intramural hematoma, ileal duplication, lymphosarcoma, inspissated meconium associated with cystic fibrosis and Henoch-Schönlein purpura. Fewer than 10 percent of children with intussusception have a leading point; its occurrence is even less common, perhaps 2 percent to 3 percent, in children less than 2 years old.

The development of intussusception is associated with venous stasis in the mesenteric vessels, followed by engorgement and edema of the bowel wall that progresses to strangulation. The compromise of the vasculature, which results from the intussusception, produces colicky pain, reflex vomiting and bloody stools. The so-called currant-jelly stool is pathognomonic of intussusception. At times, a mass may be palpated at some point along the course of the colon; this mass represents the intussusception, bowel inside bowel, with its consequent edema and trailing mesentery and vessels. An astute clinician may detect an emptiness in the right lower quadrant that reflects the movement of

*Assistant Professor of Pediatrics and Community Medicine.

Kenneth Miller, MD (Children's Hospital and Health Center, San Diego) and David Edwards, MD (University of California Medical Center, San Diego) have granted permission to use the radiographs in this article.

Reprint requests to: Martin Stein, MD, Director, Ambulatory Pediatrics, University of California Medical Center, Pediatric Primary Care Center, P.O. Box 3548, San Diego, CA 92103.

†The WESTERN JOURNAL's style regarding eponyms is that they are not written in the possessive form; therefore, Graves disease, Ewing sarcoma and Paget disease. An explanation may be found on page 78 of the July 1978 issue.

the distal ileum and cecum away from their usual position. Eventually, if this process is allowed to continue, intestinal obstruction will ensue. If gangrene sets in, perforation and peritonitis will develop.

The goal of the primary pediatrician is to diagnose intussusception at an early stage in the disease process when reduction by barium enema may be possible, thereby permitting the avoidance of operative intervention. Several recent cases provide the framework for discussion in this Specialty Conference.

ANDRE RASZYNSKI, MD:* CASE 1. A 5-month-old girl was brought to her pediatrician with a history of occasional vomiting and diarrhea and episodes of crying and irritability over a 24-hour period. She had been in excellent health before this episode. On physical examination the infant was noted to be afebrile, well-hydrated and relaxed. The abdomen was soft and nontender. A diagnosis of acute gastroenteritis was made, and the parents were advised to provide a clear liquid diet and to make a follow-up phone call. As the infant and parents were leaving the pediatrician's office, the patient developed a sharp cry and opisthotonic posturing. Immediate examination included a rectal examination which yielded an explosive currant-jelly stool. When the baby quieted, the abdomen was again soft and without palpable mass.

At this time, the pediatrician suspected intussusception. A plain roentgenogram of the abdomen showed a density in the area of the transverse colon. A barium enema documented an intussusception in the distal transverse colon. Complete reduction of the intussusception was accomplished by means of hydrostatic pressure with barium. The barium was followed through the colon and into the small bowel. A follow-up scout film six hours postreduction did not show any evidence of obstruction or recurrence.

DR. STEIN: The acute onset of vomiting, colicky abdominal pain and rectal bleeding in a previously healthy infant represents the cardinal symptomatology of intussusception. Although all three symptoms were eventually evident in this patient, it was logical that the physician initially made a diagnosis of gastroenteritis. It is not typical, but the patient with acute intussusception may appear very well between episodes of colic. More often the infant is listless and apathetic. Episodes of pain usually recur at 10- to 30-minute intervals. Fortunately, this patient's colic, severe enough to induce posturing, recurred before departure from the physician's office, and a second examination was carried out. The importance of the rectal examination, which may yield a bloody stool, is pointed up by this case. In most children with intussusception, an abdominal mass may be palpated, but it may not. The absence of a palpable mass may delay the diagnosis unless the clinician is suspicious of intussusception.

CASE 2. A 15-month-old boy presented to the hospital after a 24-hour history of vomiting, anorexia and intermittent abdominal pain. There had been no respiratory symptoms, fever, trauma or unusual ingestion. He had been in excellent health before this illness. On physical examination he was noted to be comfortable but quiet between episodes of abdominal pain that occurred every three minutes. During these episodes he would flex his hips, moving his thighs to his abdomen, and cry in pain. The temperature was 36.6°C (98°F). There was a mobile nontender right periumbilical mass that measured 2 cm by 5 cm. Rectal examination yielded bloody, mucoid stool. A plain film of the abdomen showed, in the middle of the lower abdomen, a single dilated loop of small bowel without an air-fluid level. There was a suggestion of a mass in the right upper quadrant.

The history and physical and roentgenographic findings suggested intussusception, and a barium enema showed an intussusception in the midportion of the transverse colon. Attempted reduction by hydrostatic pressure moved the mass only to the ascending colon. The child was taken immediately to surgery, and an ileocecal intussusception was found in which approximately 5 cm of ileum extended the length of the ascending colon to the hepatic flexure. Reduction was accomplished without difficulty by means of gentle pressure. The surgeon noted several large (3 cm by 5 cm) mesenteric nodes. An appendectomy was done. The child did well postoperatively.

This case was typical of childhood intussusception in that (1) it occurred in a previously healthy infant (approximately 75 percent of cases are in children less than 2 years old); (2) the patient was a boy (it affects males more frequently than females by a ratio of 3:2); (3) vomiting and colicky abdominal pain were the presenting symptoms; (4) a mass was palpated in the right upper quadrant (abdominal masses have been discovered

*Pediatrician in private practice, San Diego.

by palpation in some 20 percent to 90 percent of patients in reported cases[1,2]).

Reduction by barium enema was not successful. In general, when symptoms last longer than 24 hours the chance for complete reduction by enema is decreased. However, in most patients with intussusception, barium enema reduction, with a maximum of 3 feet of hydrostatic pressure, should be attempted. With this pressure limit, Ravitch has pointed out that gangrenous bowel will not reduce and perforation will not occur.[1]

At surgical operation large mesenteric nodes were observed. Although mesenteric adenitis may be secondary to a viral infection, attempts to document acute viral illness, especially advenovirus infection, in patients with mesenteric adenitis and intussusception have been equivocal.

WILLIAM NYHAN, MD, PHD:* It is of interest to remember that after 24 hours of symptoms, this child did not produce a bloody stool. It was only in association with the rectal examination that a bloody, mucoid stool was identified. Bloody stools are more common in children less than 2 years old than they are in older children with intussusception, but of course this child was less than 2. Following the rectal examination, he passed a bright red stool with mucus. This is worth emphasis because in my view this is the classic currant-jelly stool. In these patients enormous amounts of mucus are produced. They may pass pure mucus per rectum and I have even seen patients with intussusception produce mucus in vomitus and by stomach tube. The so-called currant-jelly stool is an admixture of blood and mucus, and usually it is an imperfect streaky mixture, which no one would really mistake for jelly.

DR. STEIN: CASE 3. An 8-month-old boy was in excellent general health until the morning of admission. Upon awakening, he had a dark brown, diarrheal stool and started crying intermittently. He drank 8 oz of milk but then began vomiting. His pediatrician father stated that it appeared his son was experiencing abdominal pain. He became progressively more lethargic and appeared gravely ill. On physical examination he was afebrile. He appeared seriously ill and was described as septic in appearance. He had alternating periods of lethargy and crying every three to five minutes. When crying, he would flex his hips and knees, flail his arms and arch his back. The blood pressure was 100/60 mm of mercury. His neck was supple. There were no local areas of infection. Initial abdominal examination showed no abnormalities. The admitting diagnosis was sepsis. A lumbar puncture was normal. The leukocyte count was 17,600 per cu mm, with a left shift. Analysis of urine produced normal findings. Rectal examination yielded a minimal amount of guaiac-positive stool.

A subsequent examiner noted a firm, discrete, tubular mass in the right upper quadrant in an otherwise soft, nontender abdomen. A barium enema showed at the midtransverse colon a coiled spring appearance that was consistent with intussusception. Partial reduction by hydrostatic pressure moved the mass to the cecum. At surgical operation an ileoileal-colic intussusception was found at the midpoint of the ascending colon. It was reduced by gentle digital pressure. An appendectomy was done, and the child had an uneventful postoperative course.

This infant's course is an example of the profound lethargy and prostration that may be associated with intussusception at an early stage in the disease process. His initial appearance suggested shock, sepsis or meningitis. A normal blood pressure and normal findings on prompt examination of the cerebrospinal fluid suggested an alternative diagnosis.

It is characteristic of intussusception, especially in infants, to find a flaccid, exhausted patient between the attacks of abdominal colic. The abdomen is typically not distended, and tenderness may be absent or localized to the mass. A diagnosis of intussusception may be delayed until an abdominal mass is palpated. When the third examiner located a right upper quadrant mass in this patient, a barium enema was performed in order to substantiate the diagnosis. A high index of suspicion of intussusception in a young patient with vomiting and intermittent abdominal pain, coupled with careful examinations of the abdomen, will clarify the diagnosis. Occult blood in the stools or grossly bloody stools are helpful observations.

In young children, a barium enema will completely reduce the intussusception in approximately 75 percent of patients. A short duration of symptoms is said to be associated with a high reduction rate by barium enema. In this patient, an ileoileal leading point was found invaginating into the ascending colon, as opposed to the more typical ileocolic form of intussusception. Perhaps

*Professor and Chairman, Department of Pediatrics.

this anatomic variant prevented complete reduction by barium enema.

This patient did not have currant-jelly stools. He had only brown diarrheal stool that contained occult blood. The finding of normal stool on rectal examination, free of gross or occult blood, argues against the diagnosis of intussusception in patients in whom symptoms have been present a few hours.[3]

CASE 4. A 9-month-old boy was in good health until two days before admission when a mild cough, vomiting, constipation and intermittent episodes of a high-pitched cry associated with arching of his back, developed. He was admitted to a local hospital with an admitting diagnosis of an acute neurologic disease—meningitis, encephalitis or subdural hematoma. Lumbar puncture showed clear acellular cerebrospinal fluid, with normal concentrations of protein and glucose. A nurse reported brief episodes of lapsing into coma, followed by high-pitched crying with arching of the back. The infant was transferred to University Hospital, where episodes of screaming and arching of the back were observed. Physical examination showed a temperature of 37°C (98.6°F) and abdominal fullness in the right upper quadrant, with mild tenderness and guarding. There was no clearly definable mass. Although the child had not passed a stool in two days, rectal examination was followed by a currant-jelly stool.

A plain roentgenogram of the abdomen showed multiple dilated loops of small bowel without dilatation of the colon, consistent with small-bowel obstruction. A barium enema defined an intussusception of the midtransverse colon, which was reduced only partially to the cecum after two attempts with hydrostatic pressure.

At surgical operation, the distal 15 cm of the terminal ileum was found to have invaginated up the ascending colon to the hepatic flexure. Following surgical reduction, a firm, spherical 2-cm mass on the antimesenteric wall of the ileum was seen to act as a leading point. The mass was removed by wedge resection. Pathological examination showed a fluid-filled cystic structure lined by simple columnar epithelium. The histology was that of an enterogenous cyst, an inclusion cyst of the small bowel. The patient did well postoperatively.

This infant's disease was unusual in that an anatomical leading point was the cause for the intussusception. In children less than 2 years old, fewer than 3 percent of the patients have leading points. Meckel diverticulum is the most common of these.

The presenting findings in this patient suggested an acute neurological disorder rather than an abdominal emergency. Later in the course, fullness in the right upper quadrant, as opposed to a discrete mass, was found. The plain film of the abdomen was helpful in suggesting an obstructive process.

CASE 5. The patient, a 6-year-old boy, was in excellent health when he awoke at 2 A.M. complaining of severe periumbilical pain. The pain was characterized as intermittent. Every two to three minutes he would double over with pain, which migrated to the lower abdomen over the next few hours. The pain lasted approximately a minute. At home, he passed a large, bright red, bloody stool shortly after the onset of pain. He vomited several times.

On physical examination, six hours after the onset of pain, the child was noted to be afebrile and normotensive. His abdomen was diffusely tender, especially in the lower quadrants. There was no palpable mass. Auscultation showed intermittent rushes and tingling sounds corresponding with the episodes of colic. On rectal examination, a 2-cm mass, extrinsic to the rectum, was palpated at the tip of the examining finger. A guaiac-positive stool was found.

The leukocyte count was 17,500 per cu mm, with a left shift. A plain film of the abdomen showed a small-bowel obstructive pattern. The barium enema defined a multilobulated mass in the cecum consistent with an ileocolic intussusception. Reduction by hydrostatic pressure was unsuccessful.

At surgical operation, an ileocolic intussusception in the cecum was found with a Meckel diverticulum acting as a leading point. Following reduction, the Meckel diverticulum was resected and an appendectomy was done. The patient did well.

This case illustrates the most common source of a leading point, a Meckel diverticulum, associated with intussusception. Although case 4 was an example of an associated leading point in a younger child, this case is more characteristic in that older children are more likely to have a leading point than are patients less than 2 years old.[4]

Although a mass was not found on abdominal examination, the discovery of a mass on rectal examination, along with the history of abdominal colic and vomiting, led to a presumptive diagnosis

of intussusception. Ravitch stated that in 90 percent of patients, an abdominal or rectal mass is palpable. In six of 152 patients, the mass was palpated on rectal examination but not on abdominal examination. In 11 of 152 patients, the mass made a dramatic presentation through the rectum.[5]

PHYSICIAN IN AUDIENCE: *If you were successful at reducing this child's intussusception by barium enema, you may have missed the opportunity of discovering the Meckel diverticulum and thereby increased the child's risk for a recurrence.*

DR. STEIN: You are correct in pointing out one pitfall of barium enema reduction. Since children more than six years old who have intussusception are at much higher risk for an associated leading point, some pediatric surgeons plan elective exploratory laparotomy following successful barium enema reduction in older patients. Most pediatric patients with intussusception are children less than 2 years of age, who are much less likely to have an associated leading point. Alternatively, you might look for a leading Meckel diverticulum, using a sodium pertechnetate Tc 99m scan,[6] following successful nonoperative reduction in older patients. However, this approach would bypass the occasional older child whose intussusception is the first clinical manifestation of an intestinal lymphosarcoma; this is important because an isolated intestinal lymphosarcoma is curable by surgical operation. Actually, this is not a significant practical problem because most intussusceptions with leading points are not reducible by barium enema.[4]

RICHARD BUCHTA, MD:* CASE 6. I recently saw an adolescent patient with an atypical form of intussusception. Because of his age, a lymphosarcoma was considered preoperatively. He was a 14-year-old boy who was admitted for evaluation of intermittent acute abdominal pain of ten days' duration. He had had an appendectomy at the age of 3 years. His father was listed as MIA (missing in action) in the Vietnam war, and the boy had been under psychiatric care from the age of 7 until he was 10 years old.

Acute abdominal pain had developed in this patient, associated with one episode of emesis three days after the government announced that no MIA's were considered to be still alive. Examination at that time showed no abnormalities. A clear liquid diet was prescribed, and the pain subsided. Three days later there was recurrence of severe cramping pain in the lower abdomen. No fever, emesis, hematochezia or diarrhea was present. Examination showed tenderness over the region of the appendiceal scar. Rebound tenderness was questionable. Results of complete blood count, determination of erythrocyte sedimentation rate and analysis of urine were normal. Pain subsided over the next few hours, and again the boy was asymptomatic for three days. Pain recurred, and a barium enema study was done. The films showed a large filling defect in the cecum with a coiled-spring appearance typical of intussusception. It could be only partially reduced; and, under fluoroscopy, there appeared to be, in the area of the appendiceal stump, a mass acting as a leading point for a colocolonic intussusception.

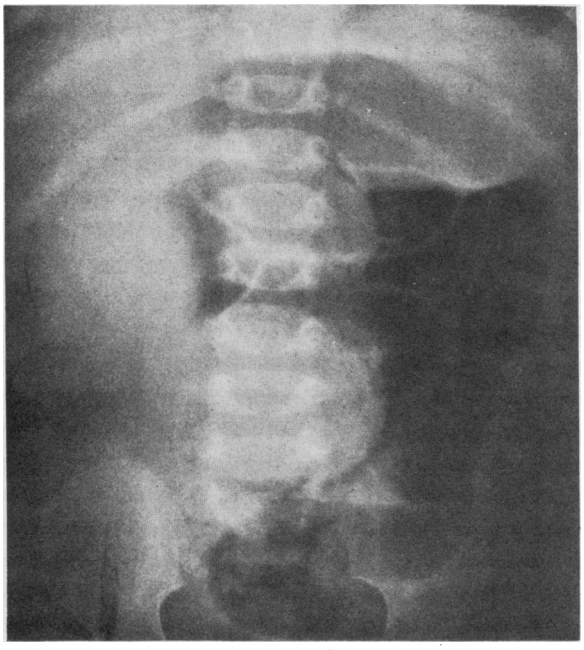

Figure 1.—Roentgenogram indicates a mass on the right side of the abdomen.

Because of the possibility of malignant tumor, a three-day bowel preparation was administered, and a right hemicolectomy was done. Pathological examination showed a 4-cm mass that almost filled the cecum. This mass consisted of a fecalith in a cavity communicating with the lumen of the cecum by means of a 1-cm opening and containing a 0.5-cm lumen. The cavity was lined by intestinal mucosa. It is postulated that this cavity was related to the earlier appendectomy. The lumen was in the area and about the same size as the normal orifice of the appendix. This must have developed suffi-

*Assistant Clinical Professor of Pediatrics.

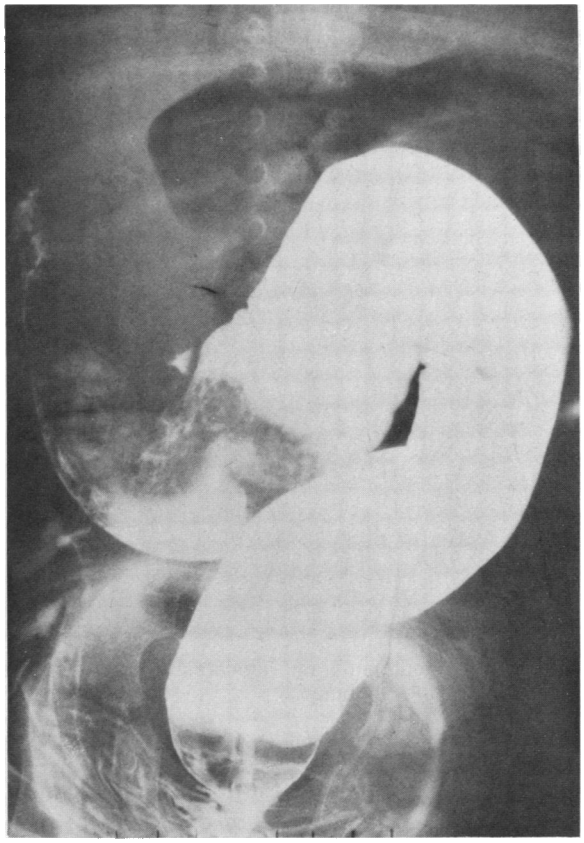

Figure 2.—The barium enema study shows an obstruction in the midtransverse colon.

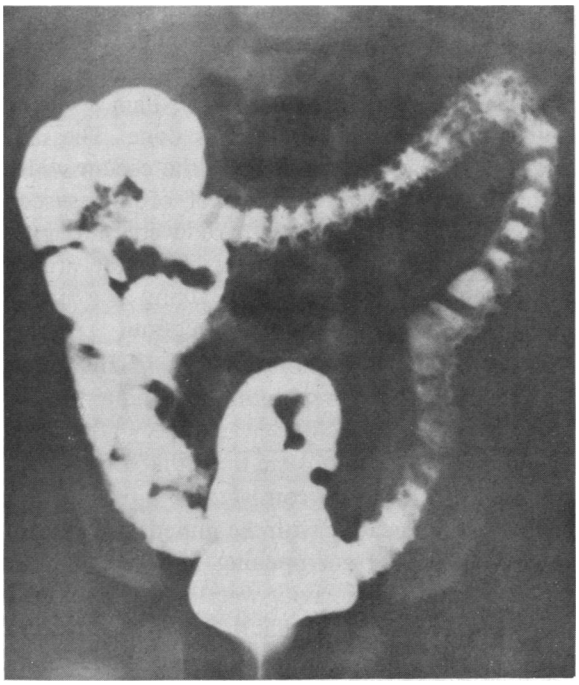

Figure 3.—The intussusception was reduced successfully by hydrostatic pressure.

cient patency to allow fecal material to enter and become trapped in the cavity, giving rise to the fecalith. A large fecalith in an intramural cyst leading to intussusception has not previously been reported.

This adolescent's prolonged course is characteristic of intussusception in older children, in whom an intussusception may remain viable and may be easily reducible as late as four days after the onset of symptoms.[7]

DR. STEIN: We have seen that roentgenograms may show intussusception in several ways. When suspicious of intussusception, a clinician may request an abdominal plain film, which may show a mass, usually on the right side (Figure 1). The mass lesion represents a portion of bowel telescoping inside another portion of bowel. The subsequent barium enema study in this patient showed an obstruction at the midtransverse colon (Figure 2), which was reducible by hydrostatic pressure (Figure 3).

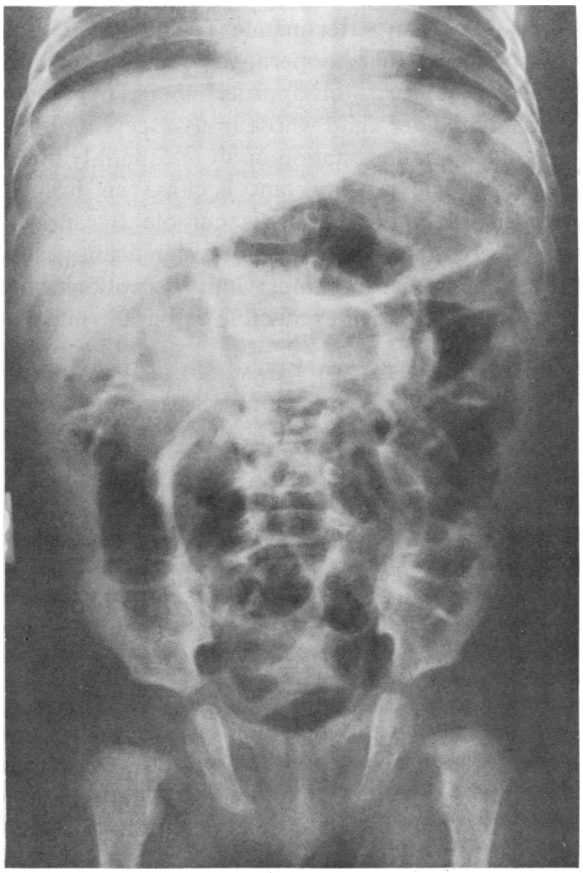

Figure 4.—The plain roentgenogram is nonspecific. There are dilated loops of bowel throughout the abdomen.

When the plain abdominal film reveals a mass or lack of bowel gas in the right lower quadrant, it may aid the clinician. However, the plain film may be normal or nonspecific (Figure 4); a barium enema study in this patient showed there to be a cecal filling defect that was consistent with intussusception (Figure 5). A more characteristic sign of intussusception is the coiled-spring effect that reflects a layering of edematous intussuscepted bowel (Figures 6A and B).

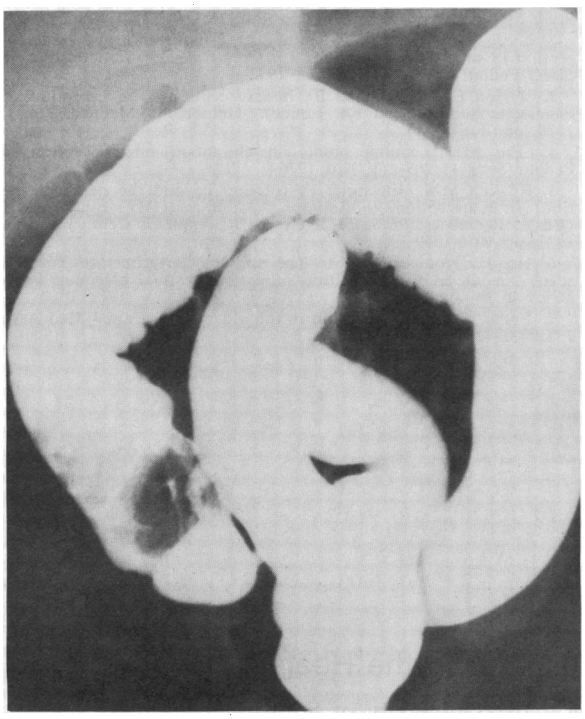

Figure 5.—The barium enema study shows a cecal filling defect which was reduced by hydrostatic pressure.

PHYSICIAN IN THE AUDIENCE: *Following successful treatment, is there a recurrence risk for intussusception?*

DR. STEIN: The recurrence rate has been reported as 2 percent. Apparently the risk of recurrence is similar in those children whose intussusception is reduced by barium enema and in those reduced at surgical operation. However, postoperative adhesions following surgical reduction may lead to obstruction. In children with a recurrence, it is particularly important to consider the possibility of an anatomical leading point. The autosomal dominant Peutz-Jeghers syndrome, in which buccal pigmentation is associated with multiple intestinal polyps, may cause recurrent intussusception with different polyps as leading points. Among the many school-age children who have recurrent abdominal pain without a documented organic cause, repeated episodes of intussusception with spontaneous reduction may be present very rarely. There have been reports of occasional spontaneous reductions that have occurred on the way from the radiology suite to the operating room.

DR. NYHAN: The diagnosis of intussusception can present a real challenge to physicians caring for children. Its manifestations and presentations have been well described as protean. At the same time, the stakes are high, because the prognosis is so very good, provided the diagnosis is made in time

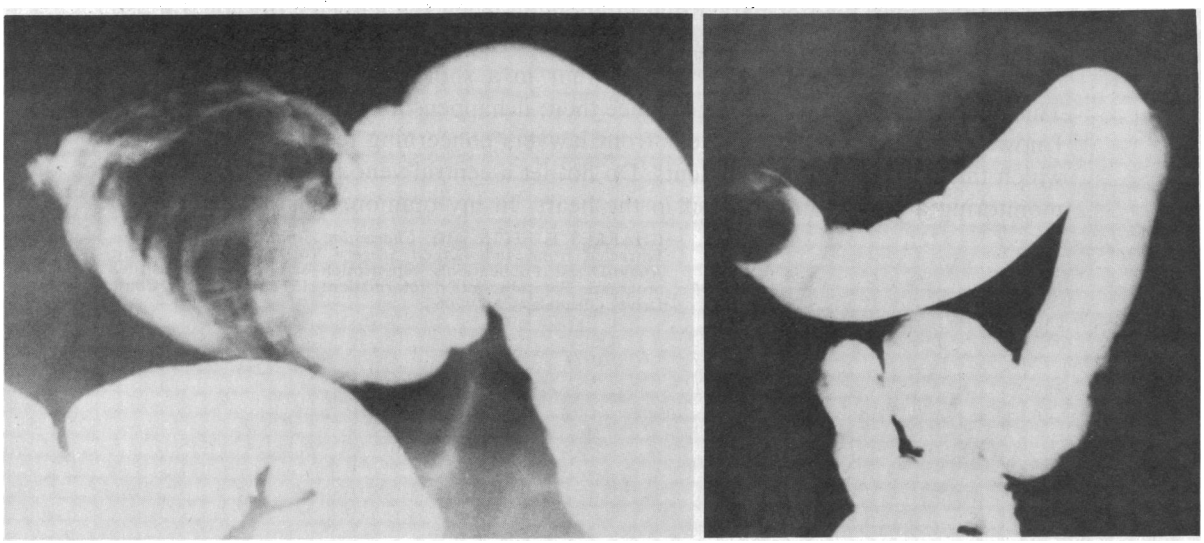

Figure 6.—A, The coiled-spring sign of intussusception. B, A different but also typical roentgenographic appearance of intussusception.

and appropriate therapy is instituted. Once the disease is suspected, proving the diagnosis is usually easy. Therefore, it is important to us to be aware of the many ways that intussusception can be present.

In this conference, we have discussed two cases in which the presentation was the typical one, with symptoms of vomiting and colicky pain in a previously healthy, usually but not always male infant less than 2 years of age. An ill-defined abdominal mass may be found, with or without bloody or currant-jelly stools. We have also discussed the typical presentation in an older child with a Meckel diverticulum as the leading point. One patient presented with diarrhea, lethargy and an appearance of overwhelming illness. On admission, he was thought most likely to have sepsis. The other infant had intermittent episodes of high-pitched crying and arching of the back and was referred with a diagnosis of acute central nervous system infection. An adolescent patient had an unusual situational presentation against a background of previous treatment for psychiatric disease. Two unusual leading points included a fluid-filled enterogenous cyst and a fecalith in a cystic structure that may have represented an appendiceal stump.

REFERENCES

1. Ravitch MM: Intussusception, *In* Mustard WT, Ravitch MM, Snyder WH, et al: Peditaric Surgery, 2nd Ed. Chicacgo, Year Book Medical Publishers, 1969, pp 914-931

2. Gierup J. Jorulf H, Livaditis A: Management of intussusception in infants and children. A survey based on 288 consecutive cases. Pediatrics 50:535-546, Oct 1972

3. Nixon HH, O'Donnell B: Intussusception—The Essentials of Paediatric Surgery, 3rd Ed. London, Heinemann Medical Books, 1976, pp 78-84

4. Ein SH: Leading points in childhood intussusception. J Pediatr Surg 11:209-211, Apr 1976

5. Ravitch MM: Intestinal obstruction beyond the neonatal period: Intussusception, *In* Rudolph AM, Barnett HL, Einhorn AH (Eds): Pediatrics, 16th Ed. New York, Appleton-Century-Crofts, 1977, pp 1056-1059

6. Ho JE Konieczny KM: The sodium pertechnetate Tc 99m scan: An aid in the evaluation of gastrointestinal bleeding. Pediatrics 56:34-40, Jul 1975

7. Fallis JC: Intussusception in the older child. Canad Med Assn J 114:38-42, Jan 1976

Hazards of the CVP Catheter in the Heart

BACK IN 1970 I found a series of patients at the University of South Carolina in whom the central venous pressure catheter was placed so that the tip of it lay in the heart. A particular case years ago was a 40-year-old man with multiple trauma . . . and that man died of pericardial tamponade when the catheter tip got trapped up in the auricle, eroded through, and fluids were given into the pericardium. We found three other cases, for a total of four in a short period at our university. This occurrence has been reported since then; it happens all over the country. And I now get at least two calls a year from lawyers concerning testimony in cases in which this has happened to patients. Do not let a central venous line that you have monitoring a patient in shock sit in the heart. In my opinion, it is dangerous.

—CHARLES T. FITTS, MD, *Charleston, South Carolina*
Extracted from *Audio-Digest Surgery* Vol. 24, No. 14, in the Audio-Digest Foundation's subscription series of tape-recorded programs. For subscription information: 1577 East Chevy Chase Drive, Glendale, CA 91206